

## Case 1/2014 - 24-Year-Old Man with Left Single Ventricle in Chronic Hypoxia

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**Clinical data:** Fatigue on mild exertion and accentuated cyanosis have been noted two years ago. He was operated at 17 days and at nine months for right and left Blalock-Taussig, respectively. The saturation remained above 85%, but Hct = 65% and Hb = 19 g/dl guided the performance of bidirectional Glenn at 19 years old. Cardiac catheterization at 20 years revealed average pulmonary pressure of 22 mmHg, which postponed the operative indication to complete the Fontan principle. He remained using aspirin, with oxygen saturation above 80%, Hct = 63%, and Hg = 20 g/dl.

**Physical examination:** Eupneic, cyanotic +, normal pulses, no jugular venous distension. Weight 58 kg, height 163 cm, BP: 90/60 mmHg, HR: 78 bpm, oxygen saturation = 83%. Palpable aorta ++ at supra sternal notch.

In precordium, ictus cordis at the 4th and 5th LICS and discrete systolic impulses in LSB. Hyperphonic heart sounds, systolic murmur, ++, rough, LSB, and tip; discrete continuous murmur at supra sternal notch. The liver was not palpable.

### Complementary tests

**Electrocardiogram** showed sinus rhythm and signs of biatrial and left ventricle overloads. P wave was peaked in I, V2-5, and extended in II, F, V5-6, with negative deflection in V1 and V2. The QRS complex was of RS morphology in V1 and Rs in V6. AQRS: +120°, AT: -60°, AP: +40°.

**Chest radiography** showed an enlarged heart of mild degree (cardiothoracic ratio: 0.54) with left long ventricular arch and also a long left medium arch. The pulmonary vascular bed was increased (Figure 1).

**Echocardiogram** (Figure 1) showed double inlet tract of single left ventricle with ventriculoarterial discordance, aorta emerging from the right rudimentary ventricle to the left and pulmonary valve atresia. The main ventricle had a diameter of 80 mm and there was moderate atrioventricular valve insufficiency to the right and mild to the left. Ventricular function was 58% by the Simpson method. The anastomosis of Blalock-Taussig type was patent on both sides and Glenn functioning. The pulmonary arteries were of appropriate size, with mild stenosis to the left.

### Keywords

Heart Defects, Congenital; Blalock-Taussig Procedure; Fontan Procedure; Pulmonary Valve Stenosis; Adult.

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**Catheterization and cardiac tomography** (Figure 2) revealed a venous vessel diverting the flow of the right superior vena cava by the innominate vein to the coronary sinus. The pulmonary artery mean pressure was 17 mmHg, and after the momentary closure of the innominate vein by balloon catheter, it increased to 19 mmHg. Pulmonary vascular resistance was 1.6 UW.

**Clinical diagnosis:** Double inlet tract of the single left ventricle, ventriculoarterial discordance with aorta to the left, pulmonary atresia, bilateral Blalock-Taussig, bidirectional Glenn, pulmonary artery stenosis to the left, atrioventricular valve insufficiency, and venovenous insufficiency to the coronary sinus, in chronic hypoxia.

**Clinical reasoning:** The clinical elements of cyanotic heart disease with decreased pulmonary blood flow and long-term hypoxemia manifest themselves as fatigue and increased hematocrit. The hyperphonic heart sounds guide to the arterial malposition with obstruction to pulmonary blood flow, compensated by systemic pulmonary anastomoses, these externalized by continuous murmur. Left ventricular overload on ECG directs to diagnosis of single left ventricle, and the emergence of atrioventricular valve insufficiency results from volume overload over time. Chest radiography suggests the aorta to the left (long middle arch).

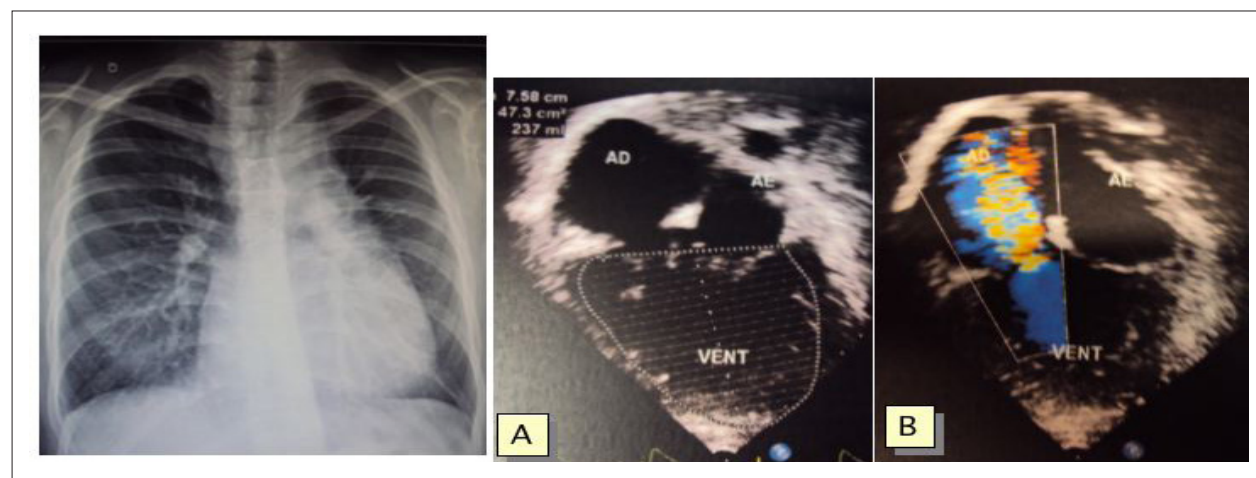
**Differential Diagnosis:** Heart diseases with rudimentary right ventricle of tricuspid atresia type are accompanied by left hemiblock. The most difficult differential diagnosis occurs in the presence of other abnormalities that are accompanied by hypoplastic right ventricle.

**Conduct:** Given the impact of long-term hypoxemia with unfavorable clinical manifestation, cavopulmonary operation was indicated, in addition to correction of atrioventricular valve insufficiency, ligation of systemic pulmonary anastomoses and venovenous vessel, and stenosis of the left pulmonary artery. The surgical risk was estimated by the marked increase in pulmonary pressure, the left ventricular dysfunction, despite of being discrete, and the other factors mentioned, in addition to the adult age. The low pulmonary resistance with high oxygen saturation constituted a mitigating factor. The surgical planning was completed with the steps previously outlined. Proceeded to the right AV valve closure, which was malformed and difficult to repair. Outer tube 20 was inserted between the inferior vena cava and right pulmonary artery, with 4-mm fenestration between the tube and the atrial cavity. The immediate evolution was favorable, with endotracheal extubation in the early hours, central venous pressure lower than 14 mmHg, and oxygen saturation above 90%.

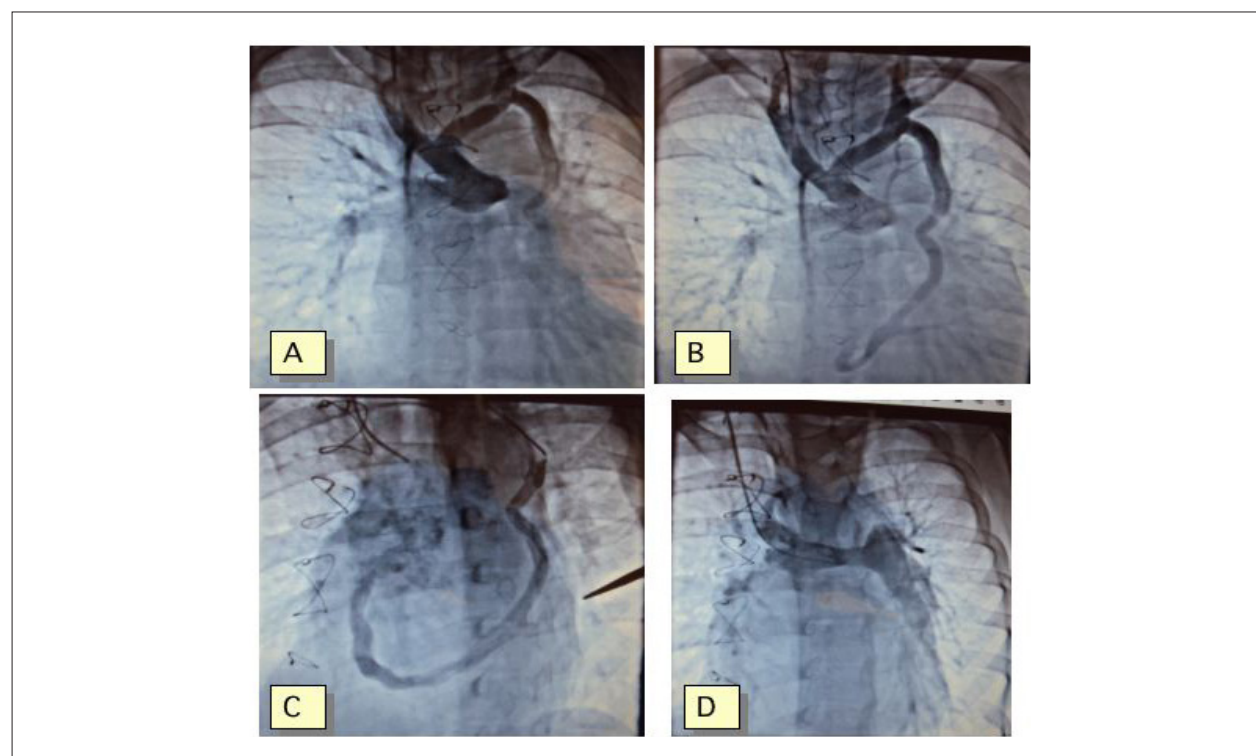
**Comments:** Although palliative, the Fontan operation, with evolution complicating factors, continues to offer good prospects provided the indication criteria are strictly observed. In the adult,

given the obligatory acquired factors in heart diseases with long-lasting overloads, the surgical risk becomes higher (10%). In this circumstance, the difficulty of the surgical indication lies on acquired aspects, such as ventricular dysfunction, anatomical lesions of the valves, in addition to high pressure in the pulmonary

artery, among others. These elements are to be counteracted with unfavorable clinical evolution resulting from representative elements of chronic hypoxia. The post-operative benefits can supersede them; therefore, clinical reasoning should prioritize elements considered reversible.



**Figure 1** - The chest X-ray shows an enlarged heart, long ventricular and long middle left arch, and increased pulmonary vascular bed. The middle arch suggests the aorta emerging from the right ventricle to the left. Four-chamber apical echocardiography highlights in A the double inlet tract of single left ventricle with two atrioventricular valves, and in B, in the same projection, moderate insufficiency of the right atrioventricular valve (mosaic of regurgitation).



**Figure 2** - Heart angiography highlights the right superior vena cava in connection with the right pulmonary artery of good diameter, and the beginning of the venovenous vessel in A, which continues bordering the heart, B, and goes into the right atrium, in C. The left pulmonary artery is of good caliber and with discrete pre-hilar narrowing in D.